

# **Parent Fact Sheet**

## **Sickle Cell Anemia (HB S/S)**

### **Cause**

Sickle cell disease is an inherited blood disorder that affects red blood cells. People with sickle cell disease have red blood cells that contain mostly hemoglobin\* S, an abnormal type of hemoglobin.

People with sickle cell conditions make a different form of hemoglobin A called hemoglobin S (S stands for sickle). Red blood cells containing mostly hemoglobin S do not live as long as normal red blood cells (normally about 16 days). They also become stiff, distorted in shape and have difficulty passing through the body's small blood vessels. When sickle-shaped cells block small blood vessels, less blood can reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes damaged. This is what causes the complications of sickle cell disease.

### **If Not Treated**

Untreated newborns often develop septicemia, an infection of the blood, and die within a few weeks of birth.

The leading cause of death in children with sickle cell disease in the United States is infection. The most troublesome germ is pneumococcus.

### **Treatment Options**

Health maintenance for patients with sickle cell disease starts with early diagnosis, preferably in the newborn period and includes penicillin prophylaxis, vaccination against pneumococcus bacteria and folic acid supplementation. Treatment of complications often includes: antibiotics, pain management, intravenous fluids, blood transfusion and surgery.

### **If Treated**

There is an extremely high mortality for sufferers under the age of five, although, better treatments are becoming available to help cope with this disease.

Treatment for the symptoms of sickle cell disease has improved over the years and many people with sickle cell disease are living beyond middle age, holding jobs, and carrying out family responsibilities.

For more information go to the following website: <http://www.newbornscreening.info>